Obstructive Biliary Tract Disease

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The techniques that have come into general use for diagnosing problems of obstructive jaundice, particularly in the past ten years, have been ultrasonography, computerized tomography, radionuclide imaging, transhepatic percutaneous cholangiography using a long thin needle, transhepatic percutaneous drainage for obstructive jaundice due to malignancy, endoscopic retrograde cannulation of the papilla (ERCP), endoscopic sphincterotomy and choledochoscopy. It is helpful to review obstructive jaundice due to gallstones from a clinical point of view and the use of the directable stone basket for the retrieval of retained stones, choledochoscopy for the same purpose using the rigid versus flexible choledochoscopes and dissolution of stones using various fluids through a T tube.

The use of dilation of the sphincter for the treatment of stenosis or stricture of the bile duct is now frowned on; rather, treatment choices are between the use of sphincteroplasty versus choledochoduodenostomy and choledochojejunostomy. Any patient with obstructive jaundice or anyone undergoing manipulation of the bile ducts should have prophylactic antibiotic therapy.

The current literature regarding treatment of cancer of the bile ducts is principally devoted to the new ideas relative to treatment of tumors of the upper third, especially the bifurcation tumors that are now being resected rather than bypassed. Tumors of the distal bile duct are still being resected by focal operations. Finally, it is now felt that early operation for congenital biliary atresia and choledochal cysts gives the best prognosis, with preoperative diagnosis now possible with the use of ultrasonography and ERCP.

THE CAUSES of extrahepatic obstruction to the bile duct include gallstones in the extrahepatic ductal system, tumors of the bile ducts and pancreas, bile duct atresia and stricture, choledochal cysts and parasites such as *Clonorchis sinesis* and *Ascaris lumbricoides*. Because jaundice due to bile duct obstruction in no way differs from that of intrahepatic cholestasis, we will begin with a very brief discussion of the traditional methods of differentiating a diagnosis of obstructive from other types of jaundice by clinical laboratory analyses that are still in use.

Obstructive jaundice begins insidiously and

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ABBREVIATIONS USED IN TEXT

ACMI = American Cystoscope Makers, Inc.
CT = computerized tomography
ERCP = endoscopic retrograde
    cholangiopancreatography
GOT = glutamic-oxaloacetic transaminase (now
    aspartate aminotransferase)
GPT = glutamic-pyruvic transaminase (now alanine
    aminotransferase
HIDA = USAN code designation for N-substituted
    iminodiacetic acid (Lidofenin)
PTC = percutaneous transhepatic cholangiography
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steadily worsens without preliminary gastrointestinal disturbances. Patients with gallstones may have had previous attacks of colic or even an episode of jaundice with the passage of a stone. Some patients with calculi may have painless jaundice. Most patients with carcinoma of the pancreas have back and epigastric pain. Cancer of the pancreas is usually preceded by pain and loss of weight and at times accompanied by steatorrhea.

Other possible diagnoses are ampullary or bile duct carcinomas. About a million new cases of cholelithiasis are discovered in the United States each year, of which about 80,000 (8 percent) have common bile duct stones. There are about 25,000 new cases of carinoma of the pancreas each year, about half of which are accompanied by jaundice, and 7,000 to 8,000 new cases of bile duct tumors. We are recognizing increasing numbers of new cases of jaundice due to parasitic infestation of the bile ducts in Asian refugees to the United States. We are also seeing strictures of the bile duct due to surgical injury at the rate of 1 or 2 per 1,000 operations on the extrahepatic biliary system.[1-3]

The liver swells considerably when the main bile ducts are obstructed, with the result that this enlargement of the liver strongly suggests mechanical obstruction rather than hepatitis or drug-induced jaundice. A palpably enlarged gallbladder suggests malignant extrahepatic obstruction rather than gallstones.

Physiologic Tests

Jaundice due to conjugated bilirubin is accompanied by bilirubinuria. This is due to reflux of conjugated bilirubin into the circulation usually due to obstruction of the bile ducts that, when complete, is accompanied by white stools devoid of bilirubin metabolites and characterized by the disappearance of urobilin from the urine. Reflux can also be caused by lesions of the liver cells due to hepatitis or cirrhosis. Liver cells continue to conjugate the bilirubin, but the excretion of conjugated bilirubin in the bile is diminished.

Serum alkaline phosphatases hydrolyze phosphoric acid esters in an alkaline medium. The levels of serum alkaline phosphatase are expressed in conventional units: their normal concentration is between 1.5 and 4 Bodansky units (BU), 3 to 15 King-Armstrong units or 20 to 85 IU per dl. The serum alkaline phosphatase levels are elevated in obstructive jaundice and infiltrative lesions of the liver. The mechanism of this increase is thought to be an increased production of phosphatases by the liver cells. One sees a moderate elevation of serum alkaline phosphatase in the course of cirrhosis or acute hepatitis. An elevated alkaline phosphatase level is not specific for liver diseases as it also occurs in association with lesions of the bones. The concentrations of the other two serum enzymes, 5-nucleotidase and leucine aminopeptidase, parallel that of the alkaline phosphatases in liver disease but their concentrations do not increase in bone disease. The isoenzymes of phosphatase can be easily identified by electrophoresis if there is doubt as to their origin.

The liver synthesizes and esterifies most cholesterol but some esterification also takes place in the plasma. The latter esterification depends on the lecithin-cholesterol-acyltransferase synthesized by the hepatocyte and released into the circulation. The serum concentration of cholesterol usually is between 180 and 250 mg per dl, with 40 percent unesterified and 60 percent esterified. The level of serum cholesterol rises in cases of obstructive jaundice because, as with the serum alkaline phosphatases, the hepatocyte produces increased quantities of this material. An elevated serum cholesterol level is not found in all cases of obstructive jaundice, and it may be elevated in some cases due to effects of drugs. The proportion of esterified cholesterol diminishes in liver failure.

Transaminases catalyze the transfer of amine groups to ketoacids. Glutamic-pyruvic transaminase (GPT), or alanine aminotransferase, and glutamic-oxaloacetic transaminase (GOT), or aspartate aminotransferase, are present in the serum. The normal values vary from laboratory to laboratory depending on method. These enzymes are present in an abundance in body tissues. The liver contains more GPT than GOT but the muscles, particularly those of the heart, contain more GOT.

The transaminases pass in large quantities into the serum in liver necrosis. Serum GPT levels are initially more elevated than GOT. The plasma levels of GOT later will be higher than those of GPT because of the prolonged survival of GOT.[1-5]

Thus, in cholestasis there is an elevation of serum concentration of conjugated bilirubin, cholesterol and alkaline phosphatase. The prothrombin time is prolonged due to reduced absorption of vitamin K, but becomes normal after parenteral administration of this vitamin.

Imaging Techniques

The simplest imaging technique for diagnosing obstructive jaundice is that of taking a plain x-ray film of the abdomen. This technique is very useful diagnostically when stones can be seen but, unfortunately, only 10 percent to 15 percent of gallstones contain enough calcium to be radiopaque. This approach is also valuable in the unusual instance where gas can be seen in the gallbladder or calcium appears in the wall of the gallbladder. A plain abdominal film should be made before any other type of study.[6]

Ultrasonography

Diagnostic ultrasonography uses sound waves that are reflected from interfaces between tissues, which produces differences in acoustic impedance. At first these instruments were capable of producing only black and white images but now can produce images with eight or more shades of grey (B mode) so that a much clearer picture of anatomic and pathologic structures can be obtained. This approach can show sonolucent bile in the gallbladder lumen and a collection of echoes in the dependent portion of the gallbladder produced by the stones, with acoustic shadows posterior to each calculus. Using ultrasonography it is possible to detect more dilated bile ducts and thus we can determine whether we are dealing with obstructive or hepatocellular jaundice. Dilatation of the extrahepatic bile ducts tends to occur in obstructive jaundice before any changes occur in the intrahepatic ducts and can be presented even before abnormalities are seen in liver function tests. A normal-sized common hepatic duct and the proximal portion of the common bile duct can be detected in oblique views anterior to the portal vein in most patients. The distal portion of the common bile duct can be seen less often. Stones are usually not seen in the enlarged distal common bile duct. Enlargement of the head of the pancreas can be

detected, but it is impossible to use this technique to differentiate between cancer and pancreatitis.[7-13] It must be remembered that normal-sized ducts may be obstructed and dilated ducts may be unobstructed (Figures 1 and 2).

Computed Tomography

Computerized tomography (CT) is an x-ray technique wherein differences in tissue density can be identified with the use of a computer. The

Figure 1.—Ultrasonography image showing the dilated common bile duct (arrow) adjacent to the portal vein in a patient with carcinoma of the distal bile duct.

Figure 2.—Ultrasonography image showing the dilated common bile duct adjacent to the portal vein (arrow) in the same patient as Figure 1, but from another angle.
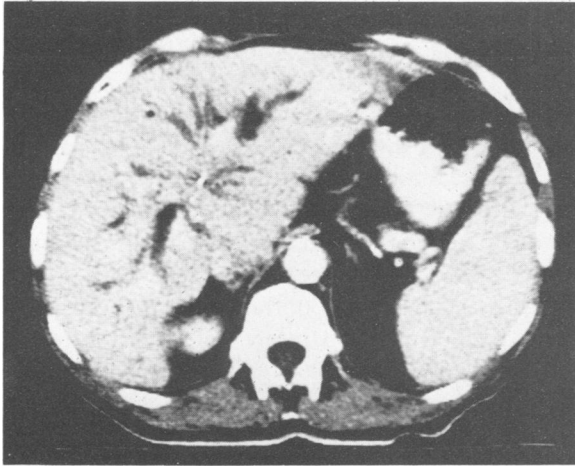
Figure 3.—CT scan image showing dilated hepatic ducts in a patient who has had resection of a tumor of the bifurcation of the hepatic ducts.

anatomic definition is much more precise than with ultrasonography but the equipment needed to carry out this type of study is very expensive and accompanied by considerable irradiation (Figure 3). This technique is more effective than ultrasonography, especially in patients afflicted with much intestinal gas and those who are extremely fat. But the absence of fat in poorly nourished patients reduces the quality of the CT image. Both ultrasonography and CT scan play an important role in the evaluation of jaundice. Identification of dilated bile ducts usually, but not always, indicates the presence of bile duct obstruction and helps distinguish between obstructive and hepatocellular jaundice. Both of these techniques can be carried out in instances where oral and intravenous methods of cholangiography are useless. Most authorities prefer to use ultrasonography first, using computed tomography when inadequate images have been obtained by the other technique.[14,15] The fact that a CT scan study costs about four times that of an ultrasound study is another point in favor of ultrasonography.

Radionuclide Imaging

An important new tool for functional and anatomic imaging of the liver and biliary tree, especially in jaundiced patients, has been technetium 99m-labeled N-substituted iminodiacetic acid (Lidofenin, or HIDA). This substance is rapidly taken up by the liver and excreted in the bile ducts in sufficient concentrations so that the gallbladder and bile ducts can be seen as it accumulates there. One can evaluate the pickup of this substance in the liver, excretion by the bile

ducts and accumulation in the gallbladder and its drainage into the intestine. It is possible to identify the gallbladder and bile ducts even in the presence of impaired liver function and serum bilirubin concentrations up to 7 to 8 mg per dl. This substance has a relatively short half-life, absence of beta emission and an ideal photon peak so that it can be given safely in high enough doses to obtain good images in a short time. The tech-
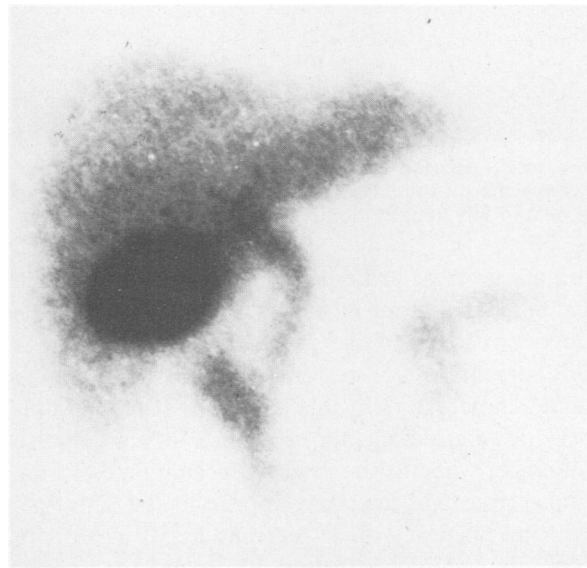

Figure 4.—Normal HIDA (Lidofenin) scan showing radioactive uptake appearing not only in the biliary tract but also in the duodenum.
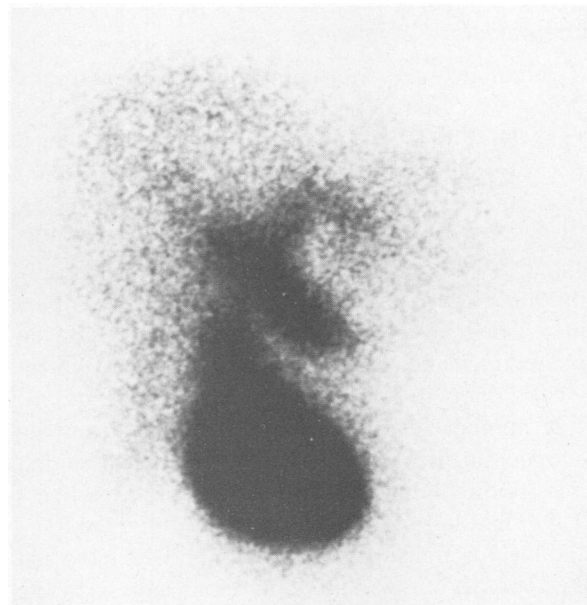

Figure 5.—HIDA (Lidofenin) scan showing an obstructed distal bile duct and gallbladder with no radioactive uptake seen in the duodenum.
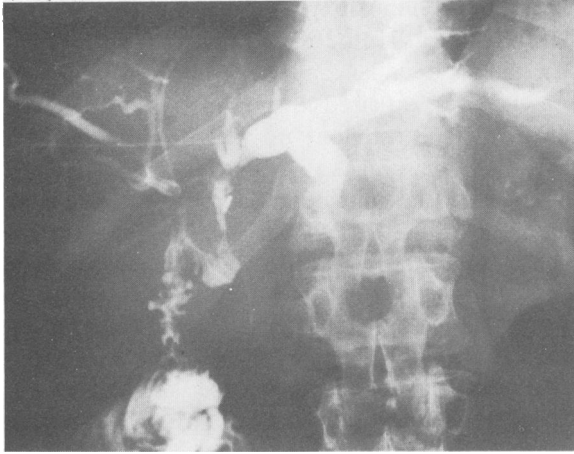
Figure 6.—Photograph of a percutaneous cholangiogram done on the same patient as in Figure 3, showing a dilated left hepatic duct.

nique's greatest value is in the evaluation of acute gallbladder attacks in which an image of the bile ducts appears and the gallbladder does not. It is 90 percent to 95 percent accurate in detecting acute cholecystitis in this situation. It also is useful in determining the presence of obstructive jaundice, wherein the material accumulates in the biliary tree and does not appear in the intestine, with an accuracy of 30 percent to 50 percent in four hours (Figures 4 and 5). In biliary atresia, it does not appear outside of the liver and a large image of the choledochus appears in choledochal cysts.[15-18]

Transhepatic Cholangiography

Percutaneous transhepatic cholangiography (PTC) was first carried out by Huard and Do-Xuan-Hop in 1937. Over the next 35 to 40 years it was rarely done because of the frequent complications of bile peritonitis, cholangitis and sepsis. Carter and Saypol[19] in 1952 placed a No. 17 spinal needle in the hepatic ducts, took x-ray films and used this needle as a drain. Ten years later, Arner and associates[20] were able to use an image intensifier so that even nondilated intrahepatic ducts in many situations can be seen.

A number of other approaches have been developed since then but a modified procedure that uses a long thin needle through the entire thickness of the right lobe of the liver appears to be the most suitable for nonsurgical patients and to have the least complications. Okuda and colleagues[21] at the Chiba University of Medicine carried out a large scale study using a 23-gauge needle with an outer diameter of 0.7 mm and a length of 15 cm that could be inserted into the liver percutaneously from the right midaxillary line. It was directed from the flank toward an area slightly above the junction of the right and left hepatic ducts with a patient on an image-intensification table, parallel to the table until the tip of the needle is positioned in the hepatic parenchyma to the right of the 12th thoracic vertebra. Contrast material is then injected slowly as the needle is withdrawn under image intensification. Contrast material runs away quickly when injected into the hepatic or portal veins, but persists in the hepatic ducts, outlines the ducts and moves slowly toward the common hepatic ducts. Insertion of the needle can be repeated safely several times until the bile ducts are seen. When the bile ducts are identified, enough contrast material is injected to fill the biliary tree and take multiple x-ray films (Figure 6).

Okuda and co-workers[21] in their initial series were able to examine the ducts of 54 of 80 patients with hepatobiliary problems and nondilated ducts. Sepsis developed in 11 of the 314 patients studied and the gallbladder was punctured in one patient. In studies carried out by Elias and colleagues,[22] cholangitis developed in two patients and in one a major leak of bile into the peritoneum after percutaneous cholangiography using this needle, both of these patients having marked dilatation of the biliary system. They were somewhat more successful in defining extrahepatic rather than intrahepatic cholestasis by percutaneous cholangiography. The group with intrahepatic cholestasis could be better defined using endoscopic approaches. Juler and associates[23] used percutaneous cholangiography in 30 patients with bile duct obstruction. They observed that 40 percent had bile leakage of 5 to 500 ml at laparotomy or autopsy and nine had peritonitis. They also observed that bacteremia developed in 23 percent. This study suggested that PTC with Chiba needle had little advantage over the larger sheathed needles and that surgical standby was required in suspected cases of obstructive jaundice. These patients should be given prestudy antibiotic agents such as cephalosporins.

Percutaneous Drainage

At this point, several authors[24-28] suggested that intubation of the hepatic ducts using a combination of the percutaneous transhepatic cholangiography technique and the Seldinger angiography catheter was satisfactory and the complications were few. A PTC using a 23-gauge needle was

carried out first. The needle was removed and an 18-cm long needle with an overlying thin-wall polyethylene catheter was installed, either in the same direction into one of the right intrahepatic duct branches or using a frontal approach under fluoroscopic control into one of the intrahepatic bile ducts of the left lobe of the liver. After bile was obtained, the needle was withdrawn and the sheath catheter was left in place through which a guide wire was inserted and positioned in one of the larger bile ducts at a suitable distance from its entry into the ductal system. The catheter was then withdrawn and a Seldinger catheter with several side holes at the tip, such as a Kifa Green catheter 1.2 mm in inner diameter, was introduced over the guide wire. The guide wire was then withdrawn and the catheter advanced farther down into the common hepatic duct.

A very large number of patients have now been reported on who had PTC of this nature with subsequent external bile drainage through tubes. This technique allows recovery of liver function and improvement in a patient's general condition before radical or palliative surgical intervention and nonsurgical palliation in advanced cases of malignancy; it also prevents postcholangiography leakage from the liver. In a series from Lund, Sweden,[25] emergency operation because of bile leakage despite established drainage of the bile ducts was necessary in only 3 out of 105 patients. In a series from Philadelphia,[24] the biliary tree was successfully catheterized in 41 of 43 cases of obstructive jaundice. Both of the failures could be traced to extensive occlusion of the intrahepatic ductal system by tumor. Once drainage was established, 22 of 39 patients had considerable reductions in total serum bilirubin at the rate of 2 mg per dl a day, with improvement in symptoms. Mean total serum bilirubin level dropped from 19.9 to 4.7 mg per dl. The major complication of percutaneous transhepatic catheterization was exacerbation of a preexisting infection with bacteremia in the obstructed ductal system despite antibiotic coverage after a "golden" period of three to five days.

Recent studies published by Ferrucci and colleagues[29] from the Massachusetts General Hospital advocate a variation of the techniques originally described by Hoevels, Ring, Nakayama and their associates.[26,30,31] They point out that guide wires of adequate caliber to support catheter exchanges will not fit the Chiba needle. For this reason, standard fine needle puncture should be

followed by puncture with a special 15-cm 18-gauge sheathed cannula in a separate step. These authors have found the biliary catheter especially designed by Ring and co-workers,[30] a 40-cm long 8.3 French multiple-side-hole catheter with a tapered tip and a pigtail terminus, is most suitable for introducing into the distal duct or all the way into the duodenum. Complications occurred in 15 out of 62 patients, of which three were major. Other complications included one case of delayed subphrenic abscess and two cases of bleeding. The complications of catheter occlusion, bleeding through the catheter, unrelieved jaundice and sepsis due to dislodgement of the catheter have been discussed at some length.

A recent study[31] showed a 29 percent operative mortality in 148 severely jaundiced patients undergoing operation without prior decompression, in contrast to an 8 percent operative mortality in 69 patients undergoing preoperative transhepatic drainage. Further refinement of the above technique has made it possible to manipulate percutaneous transhepatic catheters through an obstructing lesion into the distal segment of the bile duct or the duodenum[24] to facilitate combined external bile drainage.

Based on this experience with temporary catheters, a nonsurgical technique of introducing a permanent Teflon bile duct endoprosthesis has been developed[25,26] This endoprosthesis is inserted following percutaneous transhepatic puncture and insertion of catheters into the bile duct system and can be left in place for long periods. They can be replaced if obstructed, displaced or compromised by cholangitis, all frequently occurring complications. It appears that nonsurgical insertion of a bile duct endoprosthesis may be the only palliative procedure in situations where there is a cytologically verified malignant tumor obstructing the extrahepatic bile ducts.[27] It is probably best suited for elderly patients with poor general condition, those with generalized malignant disease, those with metastatic tumor obstructing the bile ducts and those patients in whom a surgical procedure would be impossible for technical reasons.

Endoscopic Retrograde Cholangiopancreatography

Endoscopic retrograde cholangiopancreatography (ERCP) was first described in 1968 by McCune and colleagues[32] but did not become widely available until a year later when Oi and associates,[33] using a new duodenoscope, were able to cannu-

late the papilla of Vater with greater ease using a right-angle viewer rather than a straight-ahead instrument. A cannula is introduced from below, rather than above as in percutaneous cholangiography, for injection of radiopaque medium (Fig-
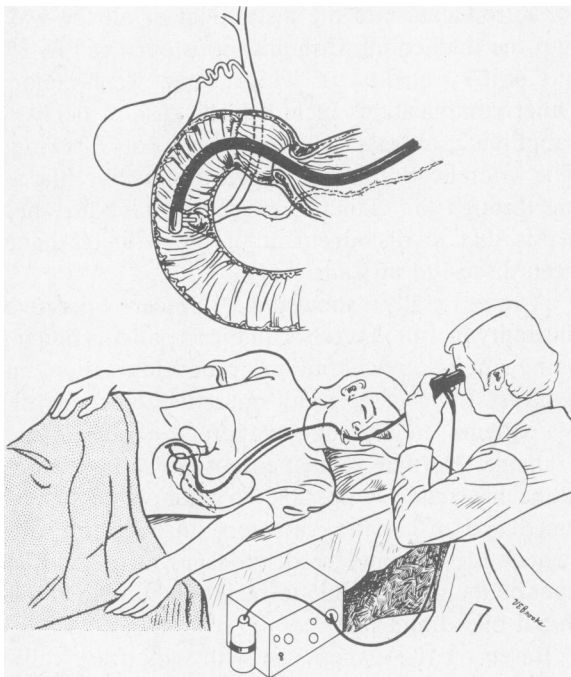

Figure 7.—Drawing showing the technique of endoscopic retrograde cholangiopancreatography (ERCP) from T. T. White and F. Silverstein (*Cancer* 1976 Jan; 37:449-461).
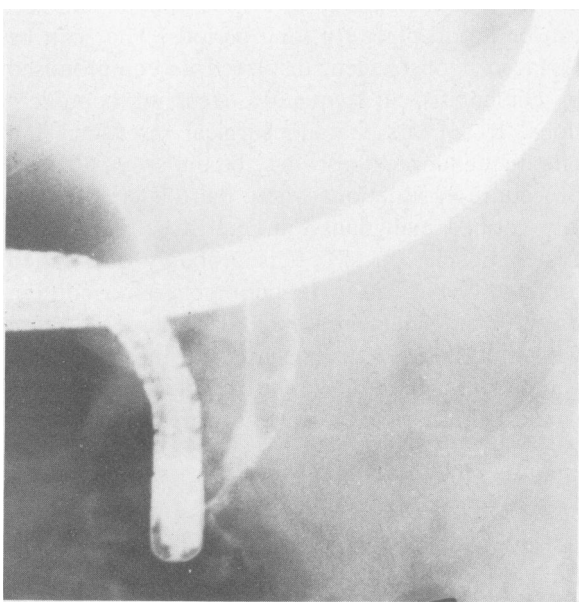

Figure 8.—Photograph taken during endoscopic retrograde cholangiopancreatography (ERCP) showing several stones in the distal bile duct.

ures 7 and 8). This is primarily an imaging procedure for obtaining good x-ray studies of the biliary system.

Sepsis and pancreatitis are the two main hazards of retrograde injection, chiefly because of the increased pressure with injection.[34,35] Pancreatitis with some degree of elevation of serum amylase level occurs in most patients in whom the pancreas is opacified, especially when overopacification of the acini occurs. Sepsis occurs primarily in patients with poor drainage of the biliary tree. Many of the patients with stasis were already infected before the injection, with sepsis and cholangitis occurring because of dissemination of bacteria into the circulation as a result of the injection.[36] When there is poor drainage of the biliary tree, injection of contrast medium should be kept to a minimum with the aim of reducing the risk of cholangitis. Otherwise, injection should continue until the biliary tree and gallbladder are filled. Tilting the patient helps to fill the hepatic tree. A number of pictures should be taken because the radiopaque medium tends to layer, which produces distortion. Calculi float up the duct. Sclerosing cholangitis, strictures, carcinoma of the bile ducts and compression of the distal bile duct by carcinoma of the pancreas or chronic pancreatitis can be seen in the cholangiograms. It may be possible to diagnose lesions high in the biliary system by inserting a brush into the biliary tree by this route to obtain specimens for cytologic examination. Cancers of the lower bile duct or papilla are easily diagnosed by visual examination at endoscopy and easily confirmed by examination of cytology or biopsy specimens.[37,38]

This is an excellent approach for differentiating obstructive from hepatocellular jaundice and for observing both jaundiced and nonjaundiced patients after biliary surgical procedure for stricture, stone or normality. Evidence obtained in this manner can serve as a guide to the treatment of an obstructed bile duct, whether by surgical or endoscopic papillotomy, surgical drainage, other types of drainage procedures or medical therapy.[38] A good review article on this subject was written in 1977 by Cotton.[36]

Cephaloridine, cefazolin sodium, and gentamicin sulfate are useful prophylactic agents when given in conjunction with these procedures. Retained dye in the biliary tract or pancreas should be drained immediately.

Endoscopic Sphincterotomy

Retained and recurrent common bile duct stones through the years have required surgical treatment. Endoscopic sphincterotomy is an extension of ERCP first described in 1974 by Kawai and co-workers[39] and Classen and Demling,[40] and several thousand such procedures have since been carried out.[41-43] The key to doing this procedure is the use of a right-angle fiberduodenoscope such as is used for ERCP through which a sphincterotome, consisting of a wire drawn through the catheter used for papillary cannulation, is inserted into the papilla (Figure 9). The distal end of the wire is fixed to the tip of the catheter so that when it is pulled, the tip of the catheter bends so that a segment of the wire separates from the catheter, producing a sawlike wire that can be used as a cautery. A mixed cutting and coagulating current is used for sphincterotomy.

The first step of the procedure is selective retrograde cannulation of the biliary tract and injection of radiopaque medium for x-ray studies. After observing the ampulla and clarifying the anatomy of the biliary system, one advances the sphincterotome into the bile duct, establishing its position by fluoroscopy. The sphincterotome is then drawn back under visual control until the proximal end of the free portion of the wire is just outside the papilla. The direction of incision should then be upwards at the 10 or 11 o'clock position when facing the papilla. The average length of the incision in the bile duct should be 17 mm.

Common bile duct stones are removed with a Dormia basket catheter. The closed catheter is advanced beyond the calculi under fluoroscopic control, then the basket is opened and an attempt made to capture the stone and remove it from the bile duct. A captured stone can be pulled out of the duodenum along with the endoscope. The overall success rate of this technique is reported to be between 90 percent and 95 percent.

The main complications have been bleeding, retroperitoneal perforation, pancreatitis, cholangitis with an impacted stone and an impacted Dormia basket, with the total incidence of complications occurring in less than 7 percent of operations done. Emergency laparotomy has been required in somewhat less than 2 percent and the overall mortality of this procedure is less than 1 percent.

Endoscopic sphincterotomy was initially used only for postcholecystectomy patients who were elderly, poor risks or in whom there were technical problems. At present, high risk patients who still have their gallbladder and in whom acute obstructive cholangitis or obstructive jaundice can be relieved before cholecystectomy are being drained in this fashion. This converts a high risk patient to one who is a candidate for an elective cholecystectomy, with well-defined biliary anatomy showing residual stones that can be flushed out. This technique, however, should be carried out only in the hands of skilled endoscopists.[41-44]

Choledochoscopy

Choledochoscopy has gradually developed since Bakeš designed a speculum in 1923 for examining the interior of the bile ducts at operation. He used an instrument somewhat like the ones used to examine the vocal cords today. Later attempts were made in the 1930's to look at the interior of the gallbladder using a cystoscope. The first choledochoscope was described by McIver in 1941[45] and manufactured by the American Cys-
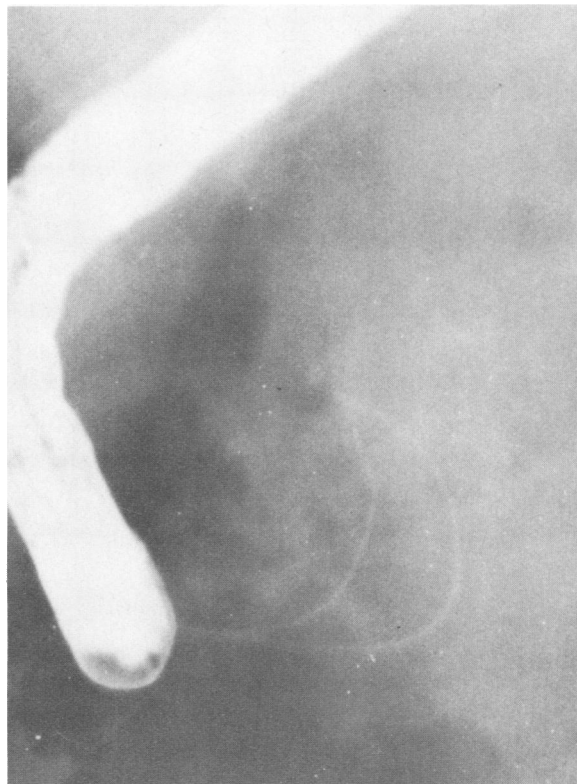

Figure 9.—Photograph taken during retrograde cholangiopancreatography showing endoscopic cannulation of the distal bile duct with the cutting wires in place for endoscopic sphincterotomy in the same patient as in Figure 8. The stones shown in Figure 8 were later successfully removed.

toscope Makers, Inc. (ACMI). This had a rather dim light with the intraductal portion of the instrument at right angles to the viewing arm. Another choledochoscope described by Wildegans and manufactured by Wolf had a 60-degree rather than a 90-degree angle. The most recent choledochoscope of this type was introduced by Storz using the Hopkins rod lens optical system, which gives a more effective optical function and uses a fiberopticlike carrier to furnish light. This rigid right-angle choledochoscope measures 3 mm in diameter and is 4 or 6 mm in length, depending on the model. An instrument guide channel can be attached for the passage of stone-retrieval instruments or biopsy forceps.

The first flexible fiberoptic choledochoscope was introduced by ACMI in 1965,[46] with an optical performance somewhat inferior to that of the rigid scopes previously introduced. More recently, several new models of flexible instruments with directable tips have been introduced by Olympus Optical Co., Fuji, Machida and ACMI[47] that can be used both at operation and through a T-tube tract. These instruments have been greatly improved over the early ACMI scope.

CHOLEDOCHOSCOPE

Objective lens
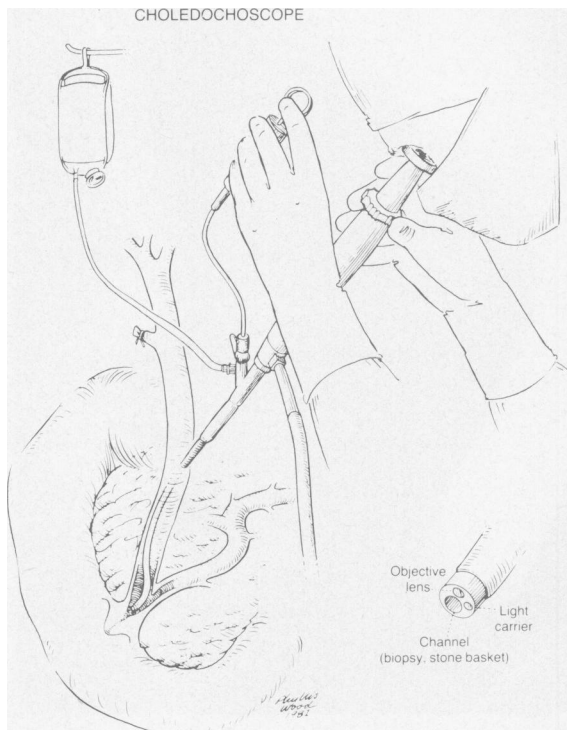
Light carrier

Channel (biopsy, stone basket)

Figure 10.—Illustration of one of the several types of available flexible endoscopes for examining the interior of the bile duct. This particular one was manufactured by American Cystoscope Makers, Inc. (ACMI).

Techniques that not only locate gallstones in the common and hepatic ducts but also reliably ensure that no stones have been left behind are essential in the treatment of benign and malignant obstructive jaundice. Intravenous cholangiography, percutaneous transhepatic cholangiography and ERCP have been used for these procedures and for operative cholangiography.[48-50] The bile duct is explored when cholangiograms show the presence of filling defects or bile duct obstruction. The common bile duct has been blindly explored for years with a number of different types of forceps and probes that also gauge the diameter of the ampulla by feel. These instruments can easily extract a stone from the duct. Flushing has been used to help remove stones. One cannot palpate extremely small stones. The rigid choledochoscopes manufactured by Storz and Wolf have the advantage of good optics and good light, together with ease of use in determining visually whether or not further sludge, stones or calculi remain in the bile duct after instrumental removal. They also are useful in taking biopsy specimens from the interior of the ducts in cases of suspected malignancy. They cannot, however, reach far up into the hepatic ducts in search of stones and cannot be used during the postoperative period through a T-tube tract. The flexible choledochoscopes vary from 5 mm (ACMI) to 6 or 7 mm (Olympus, Fuji and Machida) and can be passed down a T-tube tract following T-tube drainage of the common duct.

The operative Olympus CHF-B3 flexible choledochoscope, which has appeared recently, is a considerable improvement over the earlier flexible scopes but it is difficult to advance down the duct, as are the instruments made by ACMI, Fuji and Machida. All four types of instruments have graded amounts of flexibility and directable tips.[47,51] This type of instrument appears to be much more useful for looking high up into the liver and into the bile ducts through a T-tube tract during the postoperative period following common bile duct exploration,[47,52,53] than for routine choledochoscopy at operation where the bulk of the stones are between the bifurcation of the hepatic ducts and the ampulla (Figure 10).

The fiberoptic flexible scopes cost from $4,000 (ACMI) to $6,000 to $7,500 each (Olympus, Fuji, Machida). The rigid choledochoscopes cost $1,500 to $2,000 each. These instruments can be used only once every 24 to 72 hours because of the need for gas sterilization and aeration.

Obstructive Jaundice Due to Gallstones

Gallstone disease is thought to occur in about 10 percent of the adult population in the United States and in about 20 percent of people over the age of 40. The incidence of cholelithiasis increases progressively with aging. Thus, gallstone disease is one of our most common illnesses, with about 500,000 cholecystectomies carried out yearly for this disease. The most common serious problem of gallstone disease is that of calculi in the extrahepatic biliary ducts causing obstructive jaundice and its complications. The overall incidence of bile duct calculi in patients undergoing cholecystectomy ranges from 7 percent to 15 percent in different series, so that approximately one out of ten patients with gallstone disease has choledocholithiasis at the time of the primary biliary operation. The treatment of obstructive biliary disease with or without cholangitis or jaundice has been simplified by the addition of ultrasonography and HIDA and CT scans, ERCP and percutaneous cholangiography, which provide a clearer definition of the problems faced by clinicians and surgeons. The routine management of stone-related jaundice still involves cholecystectomy, cholangiography, common duct exploration with removal of stones and drainage of the biliary tree.

Measurements of intrabiliary pressure during operation or manometric cholangiography have been carried out in Europe for years but have only recently been added to the armamentarium of American surgeons.[54,55] These techniques show small obstructions to outflow of the biliary system whereas x-ray studies show layer deformities and filling defects caused by the presence of stones.[56,57] Flow and pressure studies are easy to do and are useful in making a diagnosis of pathology of the distal bile duct, but still need to be standardized.

The various types of choledochoscopes described above have all been developed in the last ten years and are useful for locating stones in the duct system not ordinarily found by the blind techniques used in the past. The rigid choledochoscopes are useful for more routine cases where the stones are in the common hepatic and common bile ducts. The flexible types made by ACMI, Olympus and others are especially valuable for identification of stones in more remote areas of the biliary systems, such as high in the liver.[47-53]

A useful adjunct in the postoperative period has been the possibility of removing stones through a T-tube tract that have been missed despite careful exploration. Cholic acid[58] and, more recently, Capmul (a solvent manufactured by Stokely-Van Camp, Inc. and currently under investigation) have been introduced in an effort to dissolve cholesterol stones that have been left behind after common duct exploration. Also, simple irrigation of the T tube with saline or heparin will dislodge many of these stones into the duodenum. At the same time, as these new solutions have emerged it has become possible to pass steerable catheters down into the common

T-tube in place   Directional catheter inserted   Catheter retracted, stone basket open   Stone trapped in basket   Stone removed, catheter in place
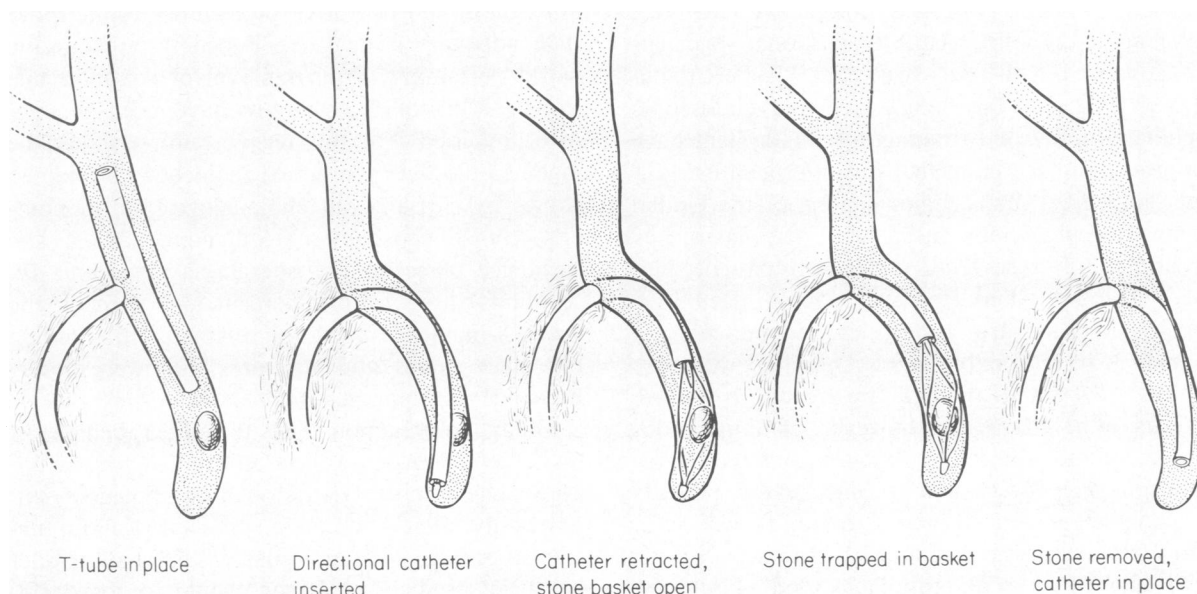
Figure 11.—Diagrammatic representation of removing stones by a directable stone basket (from White and Harrison[2]).

bile duct under fluoroscopic guidance so that stones can be trapped with a Dormia basket and pulled out through a T-tube tract to the surface (Figure 11). This technique requires that the T tube left in the common bile duct following exploration be large enough to allow passage of instruments to the bile duct. At first this was done by putting a large catheter over the outside of the vertical arm of the T tube.[59,60] T tubes are now available in which the vertical arm is four sizes larger than the part inserted into the duct, allowing a relatively small tube to be placed in the duct with a larger branch extending to the surface. A part of this technique requires that the vertical arm be run laterally so that with a patient in a supine position a radiologist can see the instrument passing through the tract without overlapping the bile duct. With this approach one must wait six to eight weeks after the operative procedure to allow formation of a solid tract that will not be damaged by instruments.

At the same time that this technique was being developed, surgeons, internists and radiologists discovered that a fiberoptic bronchoscope could be passed down through a T-tube tract into the common bile duct for extraction of stones. The special choledochoscopes described above are useful for extracting stones under direct vision in the postoperative period.[51] At present, it appears that the mechanical methods of extraction of stones in the postoperative period are more efficient than those of chemical dissolution.

The nonoperative approaches to decompression in patients with advanced obstructive jaundice, cholangitis or both constitute additional spectacular advances in the past ten years.[23-31] Percutaneous drainage of the biliary tree and endoscopic sphincterotomy and drainage of this system have progressed to the point that this approach is available in a great many major centers of the United States by physicians faced with the emergency treatment of a severely debilitated patient with overwhelming jaundice and sepsis. Definitive operative procedures can be carried out when a patient is in reasonably good general condition.

Less severely ill patients are treated as before. We have the alternative, however, of doing endoscopic sphincterotomy and drainage of the biliary tree postoperatively and in unoperated patients who do not have a T tube in place, as well as in those with stones that cannot be extracted through a T-tube tract. The approach used should be dictated by the availability of experts in the use

of these techniques. Standard techniques of cholecystectomy or drainage of the common bile ducts should be used whenever the newer techniques are not available. Whichever method is used, emergency drainage of the common bile duct for septic cholangitis should always be carried out. Orloff[61] reports that "the exact incidence of retained and recurrent bile duct stones is not known because most patients who undergo cholecystectomy with or without choledocholithotomy are rapidly restored to good health. There have been no thorough long-term postoperative studies on selected patients with gallstone disease. Most of the available data on the incidence of residual stones has been obtained during the early postoperative period and most of it has been based on cholangiography performed through a T tube in the common bile duct within two weeks of operation."[61(p404)]

The rate of retained calculi following a common bile duct exploration has been consistent, ranging from 10 percent to 13 percent of patients, while that following negative findings at choledochotomy has been substantially lower. Because these observations have been confined to the immediate postoperative period, the true incidence of retained and recurrent bile duct stones is undoubtedly higher. The incidence of residual choledocholithiasis following cholecystectomy without common bile duct exploration is unknown, but it appears to be quite low. Thus, there continues to be a prevalence of residual common bile duct stones found from several days to several years after the initial operative procedure despite more than adequate critical experience throughout the world pertaining to the removal of common bile stones. Most of these stones have been thought to be retained. For this reason, efforts should be made to discover the actual incidence of retained stones by carrying out cholangiography, manometry, choledochoscopy and other procedures. The incidence of secondary operations at various institutions varies from 2 percent to 7 percent, the greatest incidence being for retained and common bile duct stones and strictures, followed closely by cancer and pancreatitis.

Biliary obstruction due to either benign or malignant disease has been relieved by four major approaches, cholecystenterostomy, choledochoenterostomy, dilatation of the sphincter of Oddi and transduodenal sphincteroplasty. The gallbladder has been used as a drainage route for over 100 years. A recent study carried out by Dayton and

colleagues[62] showed that cholecystenterostomy failed in 5 out of 59 cases of malignancy and 4 out of 17 of benign disease for an overall failure rate of 12 percent. Our own studies suggest an even higher failure rate of nearly 20 percent.

Bakeš hoped that ampullary dilatation using a series of dilators bearing his name would allow any retained stones to pass after exploration of the common bile duct. Whereas early studies indicated that dilatation practically eliminated the possibility of residual stones, later authors such as Jones and Smith,[63] Kune and Sali[5] and Heimbach and White[64] have noted the ineffectiveness of dilatation in preventing retained and recurrent stones and have felt that transduodenal sphincteroplasty of choledochoduodenostomy were preferable to dilatation of the sphincter. Lateral choledochoduodenostomy has been reported by Rodney Smith[65] to result in a sump syndrome in over half of 250 cases reported on by another London surgeon. He has carried out transduodenal sphincteroplasty on those patients, with considerable long-term success. Kune and Sali[5] and this author have noted similar problems and solved them in the same way. These and other procedures on the biliary tract have never really been studied in a prospective fashion. It appears now, however, that it may be possible to eliminate most of the retained and residual stones appearing in the late postoperative period using endoscopic sphincterotomy. This approach has been used with increasing frequency, with the result that some recent publications suggest that endoscopic sphincterotomy is the procedure of choice for retained and residual stones.[41-44]

Strictures of the ampulla of Vater and the lower end of the common bile duct are usually sequelae of the passage of multiple stones from the biliary system into the intestine. Such strictures can be the cause of primary common bile duct stones with secondary jaundice, dilatation and cholangitis and may occur both primarily and after common duct exploration. Stenosis of the sphincter of Oddi has been described in two different ways, one suggesting that actual fibrosis of the sphincter has occurred and the other that there is inflammation of the sphincter and spasm.

The diagnosis is made frequently in Europe and South America and much less frequently in North America.[2] There is considerable ambiguity as to what this condition actually consists of. Names such as ampullary fibrosis, distal stricture, fibrosis of the sphincter, vaterian segment obstruc-

tion, phimosis of the ampulla, chronic odditis and chronic papillitis have been used. Patients with this syndrome usually have a common bile duct more than 15 mm in diameter and intermittently may have elevated serum bilirubin or serum alkaline phosphatase levels or both, especially after the injection of a stimulus to bile secretion such as secretin. The ducts are found to be dilated on ultrasound studies and percutaneous cholangiograms, the ampulla can be cannulated only with difficulty on ERCP and there are common bile duct stones present in about half of the cases. Patients with spasm or dyskinesia, however, have a normal duct size with easy cannulation by ERCP and without jaundice, chemical abnormality or stones in the bile duct. Attempts have been made in recent years to measure sphincteric pressure using balloons or perfusion with saline during ERCP cannulation. It is not clear as yet what the best treatment of spasm or dyskinesia is and whether it is necessary to go as far as in the treatment of papillary stricture and stenosis.

Cases of clear-cut stenosis or stricture of the bile duct are now usually evaluated preoperatively by percutaneous cholangiography or ERCP. Manometric cholangiography can be carried out intraoperatively and will show intrabiliary pressure of over 20 cm of water, low flow and obstruction in the distal bile duct. It will also be difficult or impossible to pass a No. 3 Bakeš dilator through the sphincter. There has been considerable discussion as to whether these secondary patients are best treated by choledochoduodenostomy, transduodenal sphincteroplasty or endoscopic sphincterotomy. Most patients are being treated with one or the other of the two former procedures, with some physicians advocating endoscopic sphincterotomy for more debilitated patients.[43,44]

If an operative procedure is to be carried out, there are certain special considerations that cause one to choose a sphincteroplasty rather than a choledochoduodenostomy (Figure 12).[63,66,67] A patient found at exploration to have not only a stricture of the ampulla but also coexistent pancreatitis that is not constricting the intrapancreatic portion of the bile duct should have a sphincteroplasty with division of the pancreatic duct sphincters.

A patient with substantial narrowing of the intrapancreatic bile duct should have a choledochoduodenostomy because the sphincteroplasty would in this situation be below the point of biliary obstruction. Sphincteroplasty is preferable
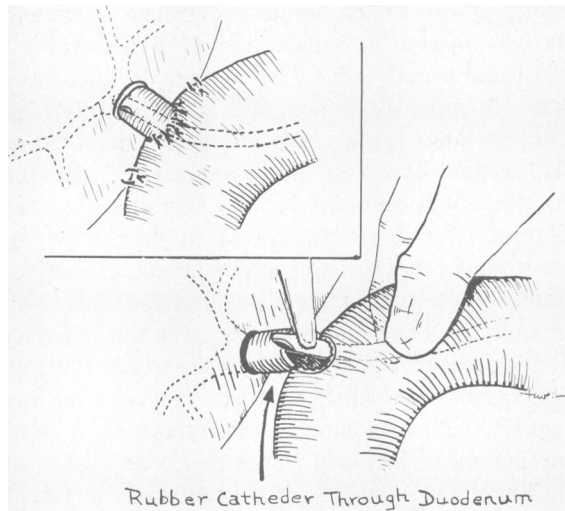
Figure 12.—Artist's drawing of a technique of choledochoduodenostomy.
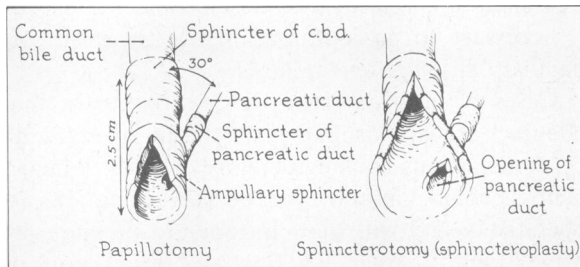

Figure 13.—Papillotomy is a quicker method of removing small stones. An incision of about 2.5 cm is required to cut the entire sphincter mechanism.

when the duodenum must be explored because of suspicion of some form of intraduodenal malignancy or because a stone is impacted in the distal end of the bile duct and cannot be retrieved otherwise (Figure 13). It is also the preferred procedure when there is associated pancreatitis without compression of the distal duct. A choledochoduodenostomy is preferable whenever the distal intrapancreatic bile duct is narrowed. We prefer to do a sphincteroplasty when the duct is less than 15 mm in diameter, a choledochoduodenostomy or sphincteroplasty when it is between 16 and 25 mm in diameter and a choledochoduodenostomy in larger ducts.[68,69]

A sphincteroplasty must be long enough to make the opening between the bile duct and the duodenum roughly the same as the largest diameter of the bile duct above. Under these circumstances one of these operations is preferred to choledochojejunostomy, because of the metabolic consequences of placing the anastomosis lower.

This approach is valuable, however, for stricture.

The most catastrophic complication of a straightforward cholecystectomy is that of accidental ligature of or injury to the common or hepatic bile duct. This usually preventable technical error is fairly rare, having occurred in 336 of 63,252 cholecystectomies done in 1973 in the state of Ohio.[70] This diagnosis should be made during or immediately after the operation but there may be no jaundice or biliary fistula and several weeks and, uncommonly, several months or years may go by before stenosis develops to the point that stricture or jaundice occurs and reoperation is required. Most such injuries are evident early by the presence of a bile fistula with accompanying jaundice, rather than jaundice alone, so that most repairs can be done during the first four to six weeks after the initial procedure. A patient who is reexplored during the first four to six weeks is often debilitated because of cholangitis and occasionally abscess formation. Under these circumstances, the biliary tract should be decompressed with catheters or a T tube and the definitive repairs postponed until infection and jaundice have subsided.

Only in rare instances where there is redundant duct that allows an end-to-end anastomosis should a biliary anastomosis be carried out. Longmire[68] reported that 11 of his 22 end-to-end anastomoses failed. The preferred approach, therefore, appears to be to anastomose the damaged bile duct to a Roux-en-Y jejunal loop both early and late (Figure 14). The anastomosis is made with either end of the bile duct to the side of the jejunum or end to end, most surgeons preferring the former. This is relatively simple when the bile duct projects sufficiently below the liver so that a good extrahepatic anastomosis can be carried out.

The anastomosis is drained internally and splinted by means of a T tube placed through the bile duct where there is enough bile duct available below the bifurcation. If the suture line is closer to the junction of the right and left hepatic ducts, the upper portion of the tube can be split. Alternatively, transhepatic tubes can be placed through both the right and left hepatic ducts where the proximal portion of the tube has been brought out through the liver parenchyma and out through the abdominal wall, as described by Cameron and co-workers,[71] Praderi,[72] Saypol and Kurian[73] and Smith.[74] These approaches have worked best when a very large tube (1 cm in diameter) was passed through the damaged or strictured area but have
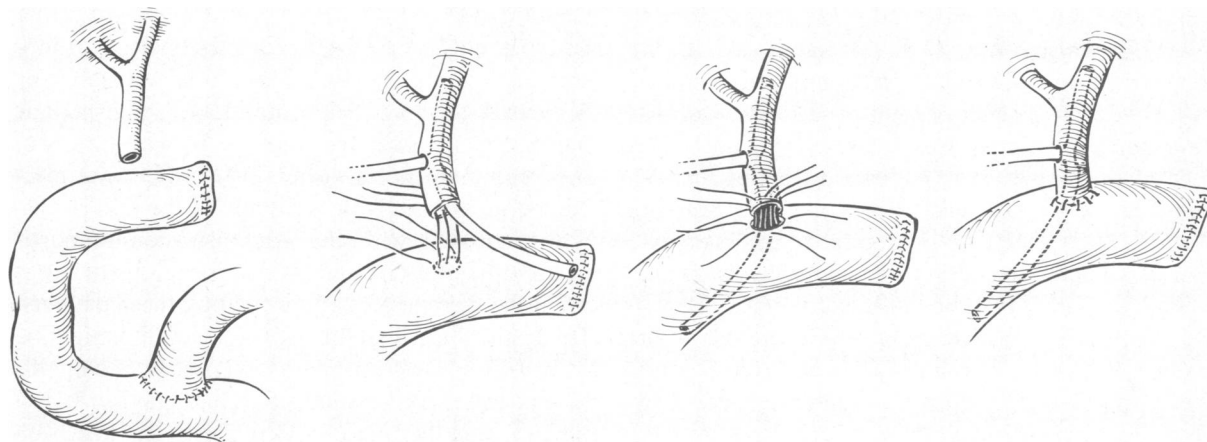
Figure 14.—Artist's drawing showing technique of choledochojejunostomy for decompression of bile duct strictures in which the damaged bile duct is anastomosed to a Roux-en-Y jejunal loop. (Reproduced with permission from White and Harrison.[4])
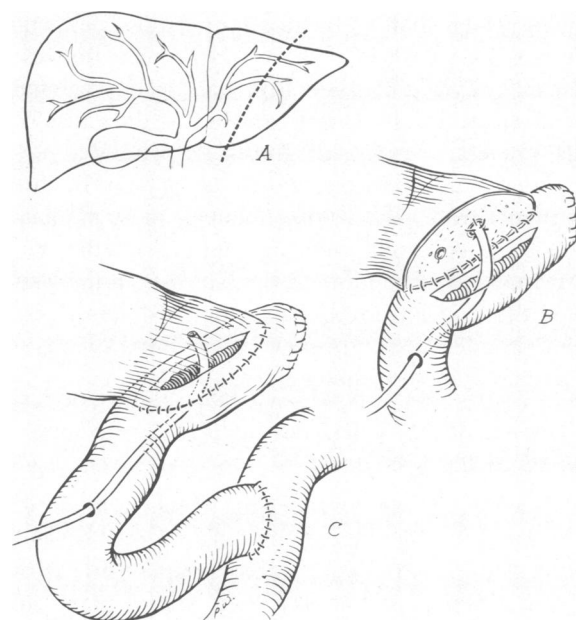

Figure 15.—The Longmire operation for high bile duct strictures. (Reproduced with permission from White and Harrison.[4])

Figure 16.—Technique of central hepatic resection for bifurcation tumors and strictures. (Reproduced with permission from Hart and White.[75])

the disadvantage of requiring daily injections to flush out debris. Further, they are often complicated by cholangitis and bleeding.

Another approach suggested by Longmire[68] is of Roux-en-Y anastomosis of one of the lateral intrahepatic ducts to a jejunal limb after a lateral segment of liver has been resected (Figure 15). We prefer removing either the entire central portion of the liver and then making an anastomosis to the several hepatic branches (Figure 16),[75] or using the technique devised by Bismuth and Corlette[76] of removing a portion of the liver to expose the main hepatic ducts away from the hilus, then making the anastomosis.

Cholangitis and Sepsis

Human bile is normally sterile but has been found to be contaminated in most patients with obstructive jaundice, common duct stones without jaundice, acute cholecystitis and patients over 60 years of age regardless of the type of pathologic

condition found.[77] Positive bile cultures are found at an increasing rate with advancing age even when only uncomplicated gallstones are present, with the result that 33 percent of patients over 70 have positive bile cultures.[78] There appears to be general agreement that patients with obstructive jaundice or anyone undergoing manipulation of the bile ducts, whether by common bile duct exploration, percutaneous cholangiography or ERCP, should have prophylactic antibiotic therapy. Cephaloridine given just before the procedure has been proved effective in reducing wound sepsis and septicemia.[79] Other effective regimens have been described using cefazolin[80,81] and gentamicin.[82] A single-dose regimen seems adequate in routine cases, rather than the three doses originally proposed by Polk and Lopez-Mayor,[83] but should obviously be continued for a long course in septic patients.

Sclerosing Cholangitis

Another type of obstructive jaundice that is confused with both primary biliary cirrhosis and carcinoma of the hepatic ducts is sclerosing cholangitis. This is an uncommon disease characterized by obliterative inflammatory fibrosis of the extrahepatic ducts, with or without involvement of intrahepatic ducts.[84,85] Patients with the following conditions are eliminated from this classification: jaundice due to drug or viral hepatitis, progressive obstructive jaundice, common duct stones, prior bile duct surgery or malignancy. All of these patients have thickening and fibrosis of the extrahepatic and sometimes the intrahepatic bile ducts. Most patients are between ages 30 and 50. Most patients are men, rather than women, as seen in primary biliary cirrhosis.

Tests of antimitochondrial antibody will show low antibody levels with sclerosing cholangitis, unlike levels found with primary biliary cirrhosis. Diagnosis has been made chiefly at surgical procedures in the past[86,87] but now can be readily made by either ERCP or transhepatic cholangiography.[88] The association of this disease with inflammatory bowel disease was reported by Warren and associates[89] and is increasingly reported to be so, with 54 percent of Wiesner and LaRusso's patients and 75 percent of Chapman and co-workers'[90] patients having associated inflammatory bowel disease.

Elevated serum copper levels are also reported with this disease, the role of which is unknown.[91] Jaundice and pruritus can be alleviated with the use of cholestyramine resin, which binds with bile and prevents its recirculation, and with corticosteroids.[92] Drainage by choledochoenterostomy or T tube is effective if appropriately established. It has been suggested recently (S. I. Schwartz, MD, oral communication, May 21, 1981) that percutaneous transhepatic drainage of the biliary system combined with the administration of corticosteroid, antibiotic and immunosuppressant drugs might be quite successful in treating these patients. It has also been suggested that colectomy might be of benefit in the treatment of sclerosing cholangitis in patients with associated inflammatory bowel disease, but the number of patients this has been done on is small. Dickson and associates[93] have suggested that penicillamine might be effective.

Most authors now think that this condition is an autoimmune reaction in the wall of the bile duct, rather than a bacterial or viral infectious disease. Association with retroperitoneal fibrosis, Riedel's thyroiditis, ulcerative colitis and regional ileitis suggest that this may indeed be the cause of the process. Positive diagnosis unfortunately requires a full-thickness biopsy specimen of the bile duct wall. Again unfortunately, most patients who have been observed for long periods die, but this may have been due to the fact that in the past only those in advanced stages of the disease were seen. Earlier cases are now being found by ERCP and percutaneous cholangiography.

Cancers of the Extrahepatic Bile Ducts and Head of the Pancreas

Cancers of the extrahepatic bile ducts occur relatively infrequently, consisting of less than 1 percent of all carcinomas and about 3 percent of gastrointestinal malignancies. A special handicap to the study of biliary cancer has been the failure of the International Classification of Diseases to distinguish between cancers of the liver and biliary system until the seventh revision in 1958. Since then about 4,500 deaths from cancers of the biliary tree have been reported yearly in the United States, as contrasted with 18,000 from pancreatic cancer. It appears that the incidence of biliary tract cancer is much higher among some racial groups than others, with Japanese having about five times the incidence of Caucasians.[94,95] The true epidemiologic makeup of this disease still remains to be clarified.

These tumors have been grouped anatomically into the following three groups according to the relative difference in prognosis of the lesions:

group 1, the upper region, including the left and right hepatic ducts and the confluence of the common hepatic duct; group 2, the common bile duct and the region of the cystic duct down to the pancreas; and group 3, the intrapancreatic portion of the common bile duct, including tumors of the ampulla of Vater.[96]

Group 3 carcinomas of the distal bile duct and pancreas may arise from the ampulla of Vater or duodenal epithelium. They may appear grossly as a polypoid growth replacing the ampulla or as an ulcer involving the same area. It is this group about which the most is known from the point of view of prognosis and treatment. They have often been grouped with carcinomas of the head of the pancreas. While they were originally treated by local resection and reimplantation into the duodenum, pancreaticoduodenectomy was first recommended for this lesion by Whipple and co-workers[97] in 1935 as a two-stage procedure. At present it is usually carried out in one stage, sometimes preceded by endoscopic sphincterotomy or by percutaneous drainage of the liver.

Pancreaticoduodenectomy is not an indicated treatment for the tumor in patients with metastases to the liver or regional lymph nodes. The mortality from this procedure has been as high as 50 percent as recently as ten years ago but shows signs of falling to about 10 percent in the hands of groups carrying out fairly large numbers of these procedures. Long-term survival over five years has been observed in around 35 percent of patients undergoing resection, which is the best outlook for survival of the various biliary tract cancers. In cases with long-term survival enough steatorrhea usually develops to require pancreatin supplements. Marginal ulcers at the site of gastric resection have developed in a number of patients and diabetes is an occasional complication.

Cases of group 2 and 3 carcinomas involving the area above the pancreas present different diagnostic and technical problems from those of the periampullary area. These have been confused with cases of gallstone disease and sclerosing cholangitis in the past, before the advent of ultrasonography, ERCP and percutaneous cholangiography.

Malignant tumors of the extrahepatic biliary tree have been noted in the past for late diagnosis in which radical cure was the exception and the prognosis was very bad. A series of 77 patients reported by Warren and colleagues[98] showed that only a third (24) had a radical procedure, 17

patients survived for more than a year and five were alive after three years. A disappointingly low survival rate has been reported in several other series.[95,99-102] Information has been scant as to the overall resectability, but some authors have reported that only a tenth to a quarter of patients are resectable at the time of laparotomy.

Jaundice has been the most important symptom of bile duct tumor and occurs quite early. The tumors are generally quite small because the lumen of the bile duct is relatively small to begin with. Other symptoms of bile duct cancer are insignificant compared with those of jaundice. Patients also have right upper quadrant abdominal pain and sometimes colic. The average duration of symptoms is two to three months. Physical examination will show an enlarged firm liver with a large gallbladder when the tumor is distal to the cystic duct or obstructing the cystic duct, causing hydrops of the gallbladder rather than Courvoisier-Terrier syndrome. Laboratory examination will show extrahepatic ductal obstruction. Ultrasonography shows dilated common hepatic and intrahepatic ducts. Both percutaneous cholangiography and retrograde cannulation of the ampulla of Vater will show the location of the tumor.

Particular attention was directed to tumors of the bifurcation of the hepatic duct by Klatskin in 1965.[103] At that time he reported 13 cases of adenocarcinoma of the hepatic duct, with advanced liver failure terminating in coma as the immediate cause of death, and he pointed out that hepatobiliary infection was the major factor in the deaths of most of these patients. He also noted that only 5 of the 13 patients had clinical evidence of metastasis at the time the diagnosis of carcinoma was made. He also pointed out that patients whose livers were decompressed lived an average of 23.3 months, as opposed to 9.9 months when the liver was not decompressed. Some authors have emphasized that these tumors should be bypassed or excised to prevent cholangitis,[104] and other publications have appeared describing methods of palliation or resection, including complete excision with end-to-end anastomosis and reconstruction of the biliary tree.[105]

The most common type of palliation in the past has been dilatation of the lesion with probes and splinting over a T tube, with resection being possible in only a relatively small number of patients, though major hepatic resection was possible in some patients.[106,107] The problem with this approach was that the T tubes became occluded

over a period and could not be replaced without reoperation. Terblanche[105] described the path of a U tube through the bile duct tumor and out through the liver, which could be replaced at will in a physician's office or in an x-ray department by simple railroading without the need for further operative procedure. This simple operative procedure resulted in the survival of some of his patients for six years or longer.

Tumors treated by the placement of U tubes extending through the liver and brought out through a Roux-en-Y loop have been offering fairly good palliation for periods of one to six years in reports from Cameron and colleagues.[71] We have found, however, that these tubes need to to be changed frequently, with considerable discomfort to the patient, and that it is possible to locally resect tumors of the hepatic ducts and bifurcation of the hepatic ducts and extend the excision up into the secondary hepatic ducts so as to achieve not only palliation but potential cures in some of these patients (Figure 16). Transhepatic tubes are used as a stent.

Attempts to excise these tumors have met with various success, with reports coming from Japan, France and the United States the past five years and articles by Corlette and Bookwalter[108] and Hart and White,[75] as well as several others.[107,109] At the present time a number of surgeons (including this author) are carrying out resection of the middle portion of the liver high up into the secondary ducts with bilateral hepaticojejunostomy, with some patients surviving without evidence of disease for as long as five years (Figure 16). Other authors are advocating resection of the right or left lobe of the liver, together with the portion of the upper bile ducts involved by tumor, and a third group of individuals is advocating removal of a portion of segment four or five in the right lobe. Also, many patients are still being treated with a U tube placed through the common and hepatic bile ducts to the anterior surface of the liver. Finally, percutaneous drainage is being used to decompress some patients in whom the diagnosis of carcinoma of the bile ducts has been made on the basis of percutaneous biopsies in tumors that appear very large on CT scan and ultrasound studies.

All of these approaches include decompression of the biliary tree, which results in fewer problems of sepsis, cholangitis and liver failure. The third, fourth and fifth approaches can be supplemented by radiation therapy, which appears to offer short-term palliation to these individuals, lengthening life for as long as six months to a year. Intraoperative radiotherapy has been advocated for patients with carcinoma at the hilus of the hepatic bile duct that involves the common hepatic artery or portal vein or that spreads to both right and left intrahepatic bile ducts (when its size does not extend beyond the field of radiation). Under these circumstances, laparotomy is carried out and external drainage is applied to relieve jaundice. A treatment cone is then positioned directly at the site of the tumor, the correct position is determined and radiotherapy is administered with an electron beam from a Betatron or linear accelerator, with a single dose of 3,000 rads and 11 to 18 meV delivered to the lesion. All patients treated thus by Iwasaki and associates[106] had slight relief of bile duct obstruction.

Another approach is that of placing radioactive gold seeds around the tumor at the time of operation. Combined with external radiotherapy, this may be of some value. The place of radiation therapy in cancer of the extrahepatic biliary system is being evaluated, both in the United States and Japan, from the point of view of curative therapy postoperatively to decrease local recurrences and increase the length and quality of survival and to treat incisional recurrences. In advanced disease management the aim is to increase the effectiveness and length of palliation and, in conjunction with chemotherapy, to further consolidate therapeutic gains made with radiotherapy alone.[110-113]

Pancreatic Cancer and Pancreatitis

Cancer of the pancreas is asymptomatic in its earliest stages; later it is evident by epigastric or back pain, jaundice and weight loss. Jaundice and a palpable gallbladder are usually the only physical findings in early cases. The same type of studies are used to diagnose obstructive jaundice on this basis as are used for the diagnosis of pancreatic cancer. A number of collaborative studies have been carried out on the diagnosis of pancreatic cancer using ultrasonography, computerized tomography, radionuclide scanning, ERCP, selective celiac and superior mesenteric angiography, duodenal drainage studies, cytologic studies and serum carcinoembryonic antigen and pancreatic oncofetal antigen assays. Most test findings have shown that a patient had either pancreatic cancer or pancreatitis. Cytologic examination with a positive result confirmed the presence of cancer

but only about a third of patients were thereby correctly diagnosed. If a negative result is taken to indicate pancreatitis, then 99 percent of all patients were diagnosed with a 75 percent accuracy.

Comparative results between ultrasound and CT scans have shown little difference. Ultrasonography is preferred on the basis of cost effectiveness but a combination may be more useful. ERCP was more accurate than duodenal drainage studies. Angiography is invasive and time-consuming and there is some difficulty with it in distinguishing between cancer and pancreatitis. Blood tests have been largely nonspecific. Therefore, gray-scale ultrasonography is probably the best noninvasive test for differentiating between pancreatic cancer and pancreatitis. CT scans are as good or superior but are more expensive. Other types of imaging procedures contribute little to this differentiation. ERCP is probably even more accurate and better at differentiating among the diagnoses of chronic pancreatitis, cancer and other lesions in this area.[114-117] The general treatment of pancreatic cancer and chronic pancreatitis is outside the realm of this paper, but jaundice in advanced cases can be treated as described earlier. The jaundice due to compression of the distal duct by chronic pancreatitis can be treated by choledochoduodenostomy or choledochojejunostomy.

Congenital Diseases of the Biliary Tract

Biliary Atresia

Congenital biliary atresia had been poorly defined up until 30 years ago. In 1953 Gross[118] reviewed 198 patients with obstructive jaundice, 183 of which were treated surgically, 26 successfully, for inspissated mucus or bile in the ducts that was treated by manipulation and irrigation of the ducts.[68] Only 12 of these patients had adequate biliary drainage achieved by duct-enteric anastomosis. Only 53 (26 percent) were relieved by medical or surgical therapy for a year or more.

There has been considerable question as to the cause of this type of lesion, whether it was due to arrested development during a solid stage, to a variety of congenital or developmental anomaly or to some type of hepatitis or cholangitis. Treatment in former years was one of delay of operation until the jaundice, if due to hepatitis, could have cleared. After the introduction in 1959 of a procedure for hepatic portoenterostomy by Kasai and associates,[119] emphasis has been on early operation, preferably in the first one to two months at a time when it is still impossible to differentiate

the hepatitis of newborn from congenital biliary atresia.

Experience with this operation has encouraged the notion that neonatal hepatitis and extrahepatic atresia are different results of a single basic process occurring immediately after birth.[120] The suggestion is that the process is probably viral in origin, causing inflammation with varying degrees of liver cell and duct epithelial injury, followed by obliteration of the bile ducts and later scarring. The suggestion is that the progressive scarring of the ductal system may be halted or even reversed by resection of the extrahepatic ducts, combined with a bile drainage procedure such as has been proposed by Professor Kasai and co-workers.[119] The possibility that many of these cases are the result of an obstructive cholangitis of varying degrees rather than an unalterable developmental anomaly gives a different point of view relative to the outcome.

It is difficult to separate the congenital from the inflammatory type of biliary atresia at the present time. Kasai attempts to enhance biliary drainage by removing all of the obstructed or potentially obstructed extrahepatic ductal system and draining the hepatic ducts into a defunctionalized loop of jejunum. Lilly,[121] A. H. Bill, MD (personal communication, April 28, 1981) and many other surgeons throughout the world have confirmed the presence of bile duct drainage from the liver following this procedure for jaundice of newborns. Kasai and associates' first patient was alive and well 18 years later. Twenty out of Kasai's 80 patients were alive without jaundice, 14 for more than two years after operation in one recent report.[119] The main problem with this procedure has been one of postoperative cholangitis that in turn may lead to cirrhosis and portal hypertension even after decompression and relief of jaundice.

Miyata and colleagues[122] reported that 15 of 51 patients were completely relieved of jaundice, portal hypertension developed in 7 of the 15 and 4 died. Most of the patients later showed transient episodes of cholangitis. Also, in a substantial number of patients who were relieved of jaundice cirrhosis of the liver later developed and they died of their disease. It is still unclear as to the dividing line between true extrahepatic atresia on a congenital basis with intrahepatic changes comparable with acquired biliary obstruction in older people, biliary hypoplasia with varying degrees of reduction in size of the extrahepatic ducts with intrahepatic cirrhosis and those with inflammatory

neonatal hepatitis. It seems logical that most cases of neonatal jaundice, excluding rare, truly developmental anomalies, are the result of an ongoing or dynamic process, possibly similar to sclerosing cholangitis in adults. The overlap between hepatobiliary pathology and the clinical course makes it very difficult to clearly differentiate those patients with inflammatory neonatal hepatitis from those with congenital atresic malformations.

Biliary Cystic Dilatation

The current classification of cystic dilatations of the intrahepatic and extrahepatic biliary systems was suggested by Alonso-Lej in 1959, discussed by Flanigan[123] in 1975. In this classification, type I cysts, or about 80 percent of the total, include dilatations of the common bile duct and common hepatic duct. Subgroups IA, IB and IC were described later. The rare type II cysts are diverticula of the common bile duct.[123] Type III cysts, or choledochoceles, are intraduodenal cystic dilatations of the terminal common bile duct that may be lined by either duodenal or bile duct mucosa. Intrahepatic cystic dilatations that present alone or in combination with extrahepatic dilatation are now called Caroli's disease.[124] Choledochal cysts with clinical signs of right upper quadrant pain, obstructive jaundice and a mass are obvious in about a third of patients and the diagnosis is obscure in the others.[125,126] About a third of reported cases come from Japan; three quarters of patients are female. A fourth of such cysts are discovered before the age of one and 60 percent before the age of ten. The diagnosis was formerly made chiefly at operation but, with the advent of ultrasound and ERCP, most such patients are now diagnosed preoperatively.[124,127]

Type II and III cysts have been and are still simply excised and managed by internal drainage. Type I cysts were formerly treated mainly by internal drainage procedures to the stomach duodenum but now are treated by excision and anastomosis to a jejunal loop.[125,128] The complications that arise in unoperated patients include cholangitis, common bile duct stone formation, common bile duct stricture, obstructive jaundice, portal hypertension, biliary cirrhosis, rupture of cysts, development of bile duct cancer and pancreatitis.[129,130] It is for this reason that external rather than internal drainage and excision with anastomosis to a Roux-en-Y loop are now carried out. This procedure seems at the present time to be the treatment of choice. It is possible, how-

ever, to leave the posterior wall of the cyst in place to avoid injury to the structure. In this case the cyst wall is opened anteriorly and the lining of the cyst in the anterior and lateral walls is removed. Reconstruction is then carried out with an hepaticojejunostomy or choledochojejunostomy.[125,129]

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